Transcatheter Stenting of Arterial Duct in Duct-Dependent Congenital Heart Disease

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INTRODUCTION

Early definitive repair and less-invasive procedures are current trends in the management of congenital heart defects (CHD). Conventional shunt surgery in duct-dependent CHD in neonates and during early infancy is associated with significant morbidity and complications, such as shunt stenosis/thrombosis, pulmonary overflow with pleural effusions, diaphragm paralysis, and distortion of pulmonary artery (PA) branches. Progress in the development of endovascular stent and implantation technique has enabled efficient alternatives to conventional surgical aortopulmonary shunts [1]. Stenting the persistent ductus arteriosus (DA) ensures its permanent patency and good pulmonary blood flow with favorable clinical response.

CASE REPORT

The complex cyanotic CHD was recognized in the first week of life when diagnosed, by echocardiography, with the pulmonary artery atresia with ventricular septal defect (PAA/VSD), an open ductus arteriosus with diameter of about 2 mm in connection with hypoplastic left pulmonary artery, and numerous major aortopulmonary collateral arteries (MAPCAs) in the right lung. The presence of large MAPCAs in the right lung enabled relatively satisfactory arterial oxygen saturation (SatO₂) of 85% and stable general condition, without the need for the introduction of prostaglandin E1. Genetic tests have confirmed 22q11.2 microdeletion.

During the follow up, at the age of three months, significant lowering of arterial SatO₂ to 55%, with a failure to thrive was identified. Heart catheterization corroborated formerly established diagnosis of PAA/VSD with DA diameter of approximately 0.5 mm at its narrowest point. Aortography clearly revealed that the left PA was supplied from DA (Figure 1), the right lung through three MAPCAs (Figure 2), and the right aortic arch with the mirror-image branching of vessels. On the basis of cardiac catheterization findings, a Cardiosurgical Board decided that an aortopulmonary shunt operation would involve a high level of risk, due to the hypoplastic left pulmonary artery and the left subclavian artery (diameter: 3 mm). Arterial duct stenting was suggested as an alternative palliative treatment.

Stenting technique: A 4-month-old female infant, weighing 3.5 kg, was re-catheterized using the trans-femoral approach, under general anesthesia. Antibiotic prophylaxis and Heparin 50 U/kg were administered at the beginning of the procedure. Aortography showed a small, 16 mm long DA extending at a very sharp angle from the left brachiocephalic artery: the narrowest point of DA was approximately 0.5 mm...
The left PA was poorly-developed at just 3 mm in diameter. After placement of the 0.014-inch coronary wire deep in the left PA, a 5 Fr guiding catheter was introduced through the DA to the left PA and, finally, the coronary stent was introduced. An 18 mm long, 4.5 mm diameter stent was chosen in order to cover the whole length of the DA (Cordis, BX VELOCITY 4.50x18 mm). After inflating the balloon catheter, we recorded a final stent diameter of 3.5 mm, which had a mild waist at the junction of DA and the left PA (Figure 3). Removal of deflated balloon from the stent was difficult, as a result of the sharp angle of the guiding catheter, but was successful without stent migration. Control aortography confirmed good stent position, covering the entire length of DA without protrusion into the aorta and with mild protrusion in the lumen of the left PA, without the risk of perforation. The flow through the stented DA was considerably higher, with a significant expansion of the entire left pulmonary vasculature (Figure 4). Arterial oximetry recorded a significant increase of the arterial saturation of 21% (from the SatO₂ of 55% before the intervention to 76% afterwards). Following the intervention, the infant had a 24-hour, continuous infusion of heparin at a dose of 20 IU/kg/hour, and commenced anti-aggregation therapy with aspirin.

After the interventional procedure, echocardiography confirmed plentiful, continuous flow through the stented DA, with a maximum pressure gradient of 46 mmHg, and a significant increase in diameter of the left PA. Further
clinical follow-ups verified an improvement in the child's growth-progress and satisfactory arterial SatO₂, with a trend of slow gradual reduction over time. One year after DA stenting, heart catheterization was repeated, in which the SatO₂ was 79%, with significant in-stent stenosis of DA (narrowest diameter: 1 mm) and excellent development of the left PA (diameter: 9 mm).

DISCUSSION

Stenting of DA represents an extraordinary technical challenge in duct-dependent CHD, since the procedure is generally performed where DA is the only source of pulmonary or systemic circulation. In these critical hemodynamic situations, even the smallest mistake or inexperience can lead to fatal consequences [2-5]. Common additional problems in the correction of duct-dependent CHD are variable morphology and spatial orientation of DA, which make the stenting procedure even more challenging. The anatomy and the spatial pattern of DA determined the optimal vascular access (femoral, axillary, carotid or trans-pulmonary) and characteristics of the stent. When choosing a stent, special attention should be paid to the length, diameter, and stent design. A stent with a larger diameter, made of thick mesh with smaller holes, allows better support of the DA wall and tends less towards causing prolapsing intraluminal tissue and consequent in-stent restenosis. On the other hand, this feature decreases the stent flexibility, which is very highly recommended in case of tortuous forms of DA. Special attention must be paid to fully covering the pulmonary end of DA with the stent, as this area has the greatest potential for constriction to the point of complete closure. Therefore, one of the most important aspects of the procedure is choosing the correct length of the stent in order to cover the entire length of DA, without protrusion into the aorta and with minimal protrusion into the PA, thus preventing constrictive reactivity of the DA terminal part.

Since the stented DA is physiologically similar to the central aortopulmonary shunt, an optimal stent diameter is 3–4 mm for neonates, because the excessive expansion of DA may produce unwanted pulmonary overflow. Another potential problem in the aftermath of the procedure is tissue proliferation with subsequent neo-intimal “in stent” stenosis and that is why children should receive continuous antiplatelet therapy. In the case of early in-stent stenosis and a significant drop of arterial saturation, it is possible to perform re-dilatation or even re-stenting of the arterial duct.

Stenting of DA, in comparison with the classical surgical aortopulmonary (AP) shunt, has many advantages: lower physical trauma, avoidance of thoracotomy and the formation of adhesions, faster recovery, and less expensive treatment [1]. However, durability of the stent in DA is limited with neo-intimal proliferation and potential thrombosis [6]. Therefore, attempts have been made to design new antiplatelet drugs or drug eluting stents that will solve these problems [7, 8]. Furthermore, there are several common complications in relation to DA stenting, including the vascular injury of entry vessels, perforation of DA or pulmonary artery, inadequate position or dislocation of the stent and worsening branch pulmonary artery stenosis [9].

In summary, the tendency of faster evolution towards the obstruction of a stent means the surgical AP shunt is no more imperfect than the non-surgical method, as both procedures are palliative and limited in duration until the appropriate age and growth of the pulmonary artery is reached [10, 11]. Considering everything, despite the existence of some technical and clinical limitations, stenting of DA can be an effective alternative to primary surgical correction in selected patients with the duct-dependent CHD.

REFERENCES

Транскатетерско стентирање отвореног артеријског дуктуса код дуктус-зависних урођених срчаних мана

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КРАТАК САДРЖАЈ
Увод Најкритичније урођене срчане мане су зависне од артеријског дуктуса (ductus arteriosus – DA) и захтевају стабилну комуникацију системског и плућног крвозвода. У почетном току се даје простагландин Е1 за постизане сталне отворености DA, а затим се хируршки креира неки од аортопулмоналних шантов. Због ризика и компликација хирурских захвата код новорођенца с критичним урођеним срчаним манама, у новије време се альтернативно у одабраним случајевима може покушати транскатетерско стентирање DA.

Приказ болесника Четворомесечном одјечету дијагностиковано је комплекска цијаногена урођена срчана мана типа атрезије плућне артерије с вентрикуларним септальним дефектом и искључивим снабдевањем левог хемиторакса из DA, док се десни хемиторакс налазио са три аортопулмоналне колатерале. Транскатетерски је без компликација постављен коронарни стент у критично стенотични и дугачки DA с крајњом димензијом лумена стента од 3,5 mm и значајно повећаним плућним протоком лево. После интервенције депе је значајно боље напредовало, а оксиметријски је забележен значајан скок артеријске сасвемености кисеоником (21%). Контролном аортографском годину дана после постављања стента утврђена је стеноза in-stent са значајно повећаним пречником леве плућне артерије и њених грана, након чега је конзилијарно одлучено да се стекли услови за даље кардиохируршке корекције.

Закључак Стентирање DA је у одабраним случајевима ефикасна альтернатива хирурским методама у примарној корекцији дуктус-зависних урођених срчаних мана.

Кључне речи: урођене срчане мане; ductus arteriosus зависне; транскатетерска интервенциона процедура; стентирање ductus arteriosus